Chapter 5

# Pre and Postoperative Management of Pediatric Patients with Congenital Heart Diseases

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Additional information is available at the end of the chapter

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**Abstract**

Stabilization during preoperative cardiac surgery especially in neonates has an important role to predict outcome for pediatric congenital heart surgery. We tried to elaborate general guidelines on how to diagnose and some anticipations for emergency treatments tailored by the type of congenital heart disease in neonates. Stabilization consists of medical treatment including emergent prostaglandin institution in some types of duct dependent lesion. The role of interventional catheterization such as patent ductus arteriosus (PDA) stent, balloon pulmonary valvotomy, etc. as modalities for stabilization before surgery was also elaborated. Some general and specific guidelines based on the type of surgeries for postoperative management were also discussed.

**Keywords:** pediatric, congenital heart disease, preoperative stabilization, postoperative management

## 1. Initial treatment of critically ill neonates with cardiac defects

In the following paragraphs of this chapter, the general features of various congenital heart defects, their clinical symptoms and treatment principles for the management of congenital heart defects that become symptomatic in neonates will be discussed. In this context, the special measures for the initial treatment of the most common defects are presented [1–8, 11].

### 1.1. Epidemiology

Based on many epidemiological studies, the actual incidence of congenital heart defects is 8–11 per 1000 live births independent of ethical background, social welfare or medical standards.

As a rule thumb nearly half of these children require any form of surgical or interventional intervention within the first year of life. With adequate management, more than 90% of these children can reach adulthood and lead a relatively normal life.

### 1.2. Symptoms

It is typical for the different congenital heart defects to present and become manifest at different times in early life. Of most importance are the very early phase when the patent ductus arteriosus closes for all ductal-dependent cardiac defects; the next important timeframe of clinical manifestation is the reduction in pulmonary vascular resistance (PVR) for defects with left-to-right shunt.

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**Note**

Typical leading symptoms for the presentation of congenital heart defects in neonates are as follows:

– Heart failure or cardiogenic shock (usually in the first or second week of life if there is left heart obstruction; for shunt defects typically not until after the drop in pulmonary resistance at the age of 2–8 weeks)

– Cyanosis

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Additional important clinical symptoms that may lead to the diagnosis of a congenital heart defect in neonates are the presence of a murmur on auscultation or any form of arrhythmia (rarely as the primary symptom of a congenital heart defect).

#### *1.2.1. Heart failure*

The characteristic clinical signs of heart failure in neonates are as follows:

- Tachypnea, dyspnea, thoracic retractions
- Rarely but possible pulmonary edema mainly due to overflow
- Tachycardia
- Hepatomegaly
- Failure to thrive, difficulty in feeding, increased sweating, abnormal sleepiness
- Pallor, prolonged capillary refill time

It is evident that the clinical symptoms of neonatal heart failure are unspecific and may be similar to the clinical symptoms of sepsis. Therefore, many neonates with heart failure are treated for suspected sepsis.

In particular, cardiac defects with left-sided obstruction (coarctation of the aorta, critical aortic stenosis, hypoplastic left heart syndrome), or typically become manifest as early as the first 1–2 weeks of life with the clinical symptoms of cardiogenic shock caused by early duct closure.

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*1.2.2. Cyanosis*

- Cyanosis is a clinical sign that becomes evident when about 5% of the hemoglobin is deoxygenated. In neonates with polycythemia (Hb >20 g/dl), this may be present even without hypoxemia, in patients with anemia this may be overseen. To avoid this, pulse oximetry at all four limbs is the standard of care.
- *Neonatal screening*: Cyanosis in a newborn should be picked up by a neonatal screening program that measures saturation by pulse oximetry at all four extremities at the first day of life. It should be standard of care that all newborns should have this screening test. If the saturation is >95% and the control 12 h later is again >95%, a cyanotic heart defect can be ruled out. If the saturation is ≤95%, additional examinations (today—ECHO as the preferred method of choice) are needed.
- *Hyperoxia test*: The hyperoxia test is a more historical test and no longer useful in the times of ECHO. It was used to try and distinguish between cardiac and pulmonary cyanosis. In patients with pulmonary cyanosis, the oxygen saturation increases markedly after oxygen is given, while oxygen saturation is usually not significantly improved in those with cyanotic heart defects. This test lacks specificity and sensitivity to rule out or confirm cardiac defects. Both pre- and postductal oxygen saturation should be measured.
- Cardiac cyanosis can be the result of either reduced lung perfusion due to a right-to-left shunt, complete venous mixing in patients with total anomalous pulmonary venous return or intracardiac mixing of systemic and pulmonary venous blood. Methemoglobinemia is another rare cause for cyanosis. In addition, cyanosis can be caused by abnormality in the central nervous system (e.g., after cerebral hemorrhage or in cases with central apnea in premature neonates, etc.)

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**Note**

All newborns should have a neonatal screening for congenital heart defects immediately after birth and 12 h later. Today ECHO is the main diagnostic tool for secondary investigation and to rule out or confirm congenital heart defects.

If the diagnosis can not be immediately confirmed by echocardiography, an attempt of treating the neonate with intravenous prostaglandin E1 is justified.

The hyperoxia test is no longer standard of care. If there is no adequate increase in oxygen saturation in a cyanosed neonate, a congenital heart defect with ductal dependent lung perfusion must be assumed.

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*1.2.3. Heart murmur*

As innocent murmurs are unlikely in this age group, a heart murmur in a neonate is always suggestive of a congenital heart defect. Usually, stenosis of the semilunar valves or AV valve insufficiency present immediately after birth due to a loud systolic murmur, the typical VSD murmur in a large VSD, for example, can often be not auscultated until after the pulmonary resistance has dropped and the pressure gradient between the left and right ventricles

increases. In addition, in many critical heart defects, there might be no indicative heart murmur at all (e.g., d-TGA simplex, coarctation of the aorta).

*1.2.4. Arrhythmias*

Arrhythmias often present in neonates without structural heart defects (i.e., atrial flutter, congenital AV block) and therefore are rarely the first symptom of a congenital heart defect. An AV block may occur in association with an l-TGA or heterotaxy syndromes. Supraventricular tachycardia due to accessory conduction pathways occurs more frequently with an Ebstein anomaly. The most common arrhythmia in a newborn is atrial flutter that requires immediate electrical cardioversion after diagnosis with adenosine but usually presents without additional defects.

### 1.3. Hemodynamic situation

The congenital heart defects that become symptomatic in neonates can be divided into five groups (**Table 1**).

| Group | Examples |
|---|---|
| Cardiac defects with ductal-dependent systemic circulation (left heart obstructions) | Critical aortic stenosis, hypoplastic left heart syndrome, interrupted aortic arch, critical coarctation of the aorta |
| Cardiac defect with ductal-dependent pulmonary circulation (right heart obstructions) | Critical pulmonary stenosis, pulmonary atresia with intact ventricular septum, pulmonary atresia with VSD, pronounced form of Tetralogy of Fallot, severe Ebstein's anomaly, tricuspid atresia with pulmonary atresia or high-grade pulmonary stenosis |
| Cardiac defects with parallel circulation | d-TGA—transposition of the great arteries |
| Cardiac defects with complete intracardiac mixing of blood | Total anomalous pulmonary venous connection, univentricular heart |
| Cardiac defects with a large left-to-right shunt symptomatic usually after 4–6 weeks | Large VSD, complete AVSD, large PDA, aortopulmonary window, Truncus arteriosus communis, univentricular hearts with unobstructed pulmonary blood flow |

**Table 1.** Classification of congenital heart defects which become symptomatic in the neonatal period.

In a ductal-dependent defect survival depends on the persistence of a patent ductus arteriosus. In general, ductal-dependent systemic circulation must be distinguished from ductal-dependent pulmonary circulation. In patients with ductal-dependent systemic circulation, there is a high-grade obstruction of the structures of the left heart (mitral valve, aortic valve, arch and coarctation). To ensure sufficient systemic perfusion, the systemic circulation must be supplied with blood from the pulmonary circulation across the patent ductus arteriosus like in utero. In ductal-dependent pulmonary circulation, there is a high-grade right heart obstruction leading to cyanosis. Perfusion of the pulmonary circulation depends mainly on the blood supply from the aorta across the patent ductus arteriosus.

In parallel circulations (d-TGA), survival depends on additional shunts (especially a sufficiently large atrial shunt) between the two circulations to enable mixing of the cyanosed and the arterialized blood.

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In cardiac defects with complete intracardiac mixing of blood, the resulting cyanosis is often only relatively mild; when excessive pulmonary blood flow due to the lack of pulmonary stenosis is often present simultaneously, this leads to pulmonary recirculation of the saturated blood. Heart failure usually develops as a result of the excessive pulmonary blood flow after 4–6 weeks.

Based on this principle, congenital heart defects with a large left-to-right shunt do not become clinically symptomatic until the age of about 4–6 weeks; at that time, the PVR has dropped and the shunt between the systemic and pulmonary circulation increases dramatically. If left-sided heart obstruction is, however, also present (e.g., VSD with CoA), the symptoms may develop as early as the first week of life.

### 1.4. Diagnostic measures

#### *1.4.1. Oxygen saturation*

As a standard of care, oxygen saturation should always be measured in children both preductally (right hand) and postductally (lower limb). If there is a defect with ductal-dependent systemic circulation, the (postductal) oxygen saturation measured in the feet may be lower than the (preductal) saturation in the right hand.

The brachiocephalic trunk branches off from the aortic arch well before the ductus arteriosus; therefore, it is safe to assume that the saturation measured in the right hand is equivalent to preductal saturation (except in the rare cases of a lusoric artery).

#### *1.4.2. Blood gas analysis (BGA)*

As metabolic acidosis is the typical finding in severe heart failure and cardiogenic shock, routine (arterial) BGA is recommended on multiple occasions. In addition, all BGA should measure lactate as surrogate of cardiac output and thereby tissue perfusion.

#### *1.4.3. Echocardiography (ECHO)*

As mentioned before, today echocardiography is the diagnostic method of choice in all patients with a presumed cardiac defect or impaired circulatory situation. ECHO allows an accurate diagnosis of all significant cardiac defects that become symptomatic in the neonatal period. As modern standard of care, every neonatal unit must have ECHO and support of a pediatric cardiologist available on a 24/7 basis.

#### *1.4.4. Hyperoxia test*

In the time of ECHO, this test has only anecdotic relevance and lacks sensitivity and specificity. This test has been used to distinguish between cardiac or pulmonary central cyanosis. After breathing 100% oxygen for a few minutes, the cyanosis disappears or should be clearly reduced in pulmonary cyanosis and there is a relevant increase in arterial partial oxygen pressure. In cardiac cyanosis, the partial oxygen pressure should remain largely unchanged as the cardiac right-to-left shunt or inadequate pulmonary perfusion cannot be compensated by the administration of oxygen.

**Note**

Today in any case, echocardiography examination is preferable to a hyperoxia test and must be available in every neonatal unit on a 24/7 basis.

*1.4.5. Pulse and blood pressure in all limbs*

All neonates with suspected cardiac defect should have a pulse and blood pressure measurement at all 4 limbs. If a difference in blood pressure between the right arm (preductal) and the lower limbs (postductal) is then manifest or the peripheral pulses in the lower limbs cannot be palpated, this is a hint and leading findings of coarctation of the aorta or an interrupted aortic arch. This examination is helpful to assess systemic circulation.

**Note**

In a large patent ductus arteriosus, there may be no difference in blood pressure between the upper and lower halves of the body even with relevant coarctation of the aorta or interrupted aortic arch. The saturations, however, vary substantially.

### 1.5. Treatment

According to the different anatomic variations and physiologic situation, there is a substantial difference in the management of these children. The treatment principles for the various groups of congenital heart defects are presented below and based on the pathophysiology, the specific treatments of the individual heart defects in the neonatal period are described in this context.

*1.5.1. Cardiac defects with ductal-dependent systemic circulation*

In ductal-dependent systemic circulation, we should reestablish the prenatal circulation. Therefore, attempts must be made to provide as much blood as possible from the pulmonary circulation to supply the systemic circulation via the patent ductus arteriosus. As a result of this strategy, a saturation of 75–85% is adequate and saturations >85% that indicate excessive pulmonary blood flow should be avoided. The treatment principles of ductal-dependent systemic circulation are as follows:

- Provide patency of the ductus arteriosus with a continuous prostaglandin E1 infusion (initial dosage 20–50 ng/kg/min, to be reduced after ECHO assessment; caveat: Apnoea).
- Lower systemic vascular resistance:
  - By reducing afterload: i.e., with sodium nitroprusside infusion
  - If catecholamines are needed: Use preferably milrinone and/or dobutamine (vasodilatation effects); avoid vasoconstrictive effects of other catecholamines (dopamine, noradrenaline)

- Increase pulmonary resistance, increase pulmonary artery pressure:
  - Avoid intubation, extubate early
  - Avoid additional oxygen
  - Aim for mild metabolic acidosis (pH 7.35)
  - Aim for mild hypoventilation ($pCO_2$ around 60 mmHg)
- In case of pulmonary edema caused by congestion and pulmonary overflow, give intravenous diuretics (1–2 mg/kg furosemide), ensure high PEEP (i.e., 10–15 cm $H_2O$) and reduce prostaglandin E1 to a minimum (e.g., 10 ng/kg/min)

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**Note**

The administration of oxygen ($SatO_2$ >85%) as a general measure and hyperventilation in children with ductal-dependent systemic circulation can lead to an acute decompensation of the hemodynamic situation.

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*1.5.2. Cardiac defects with ductal-dependent pulmonary circulation*

The situation is reversed in ductal-dependent pulmonary circulation, but the prenatal situation should be achieved too. Again a saturation of 75–85% is adequate and saturations >85% should be avoided as they indicate inadequate pulmonary perfusion. The following measures can be useful to allow more blood to flow from the systemic circulation to the pulmonary circulation via the patent ductus arteriosus if needed:

- Secure patency of the ductus arteriosus via a prostaglandin E1 infusion (initial dosage 20–50 ng/kg/min).
- Lower pulmonary vascular resistance:
  - Aim for mild metabolic alkalosis (use buffering) (pH 7.45–7.5)
  - Adjust ventilation for mild hyperventilation ($pCO_2$ around 35 mmHg)
  - Increase $FiO_2$
- Increase systemic vascular resistance and support systemic pressure by:
  - Noradrenaline infusion
  - Possibly also adrenaline infusion
  - Use volume more generously
  - Do not use dopamine
- Maintain rather high dosage of prostaglandin E1.

**Excursus: Prostaglandin**

Prostaglandin E1 is given to maintain patency or reopen the ductus arteriosus in neonates with ductal-dependent defect. Due to its short half-life, it must be administered continuously intravenous. The initial dosage in a patient with a nearly closed duct is between 50 and 100 ng/kg/min. Based on the actual effect, this dosage can be reduced gradually to a minimum of 5–10 ng/kg/min,

The most common side effects are as follows:

- Apnea (administer in readiness for intubation)
- Bradycadia
- Vasodilatation (pulmonary and systemic), hypotension
- Fever
- Edema
- (Cortical hyperostosis, periostitis only after long-term administration)

Practical tip: When a prostaglandin infusion is required, always a second venous access should be inserted to ensure safe prostaglandin administration immediately via other access if the first one is dislocated. The second access can also be used for volume substitution in case of acute hypotension.

*1.5.3. Cardiac defects with parallel circulations*

The only example of this cardiac defect is a transposition of the great arteries (d-TGA) together with its variations. The management of this situation is described in the specific treatment section.

*1.5.4. Cardiac defects with complete intracardiac mixing of blood*

The different cardiac defects (i.e., univentricular hearts) in this group are also described in the following treatment section.

*1.5.5. Cardiac defects with a large left-to-right shunt*

When the pulmonary resistance drops after the first 4–6 weeks of life, the left-to-right shunt and thereby pulmonary blood flow will increase dramatically. Consequently, heart failure may develop caused by increased volume load, which must be treated medically (diuretics, ACE inhibitors, beta blockers, possibly digoxin or cathecholamines) until corrective surgery is performed. As additional oxygen will lower the pulmonary resistance and increase excessive pulmonary blood flow, no additional oxygen should be administered.

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## 1.6. Specific treatment of the most common symptomatic heart defects in the neonatal period

*1.6.1. Left ventricle outflow tract obstruction lesion (i.e., critical aortic stenosis, coarctation of the aorta and interrupted aortic arch)*

The initial management of these children has some common features:

- Shock therapy, including intubation and ventilation (high PEEP (>10 cm $H_2O$) if there is pulmonary edema)
- Prostaglandin E1: initial dosage 50–100 ng/kg/min
- Oxygen: avoid excessive administration, use PEEP to improve oxygenation. (note: oxygen lowers pulmonary resistance and thus increases pulmonary blood flow)
- Use diuretics (furosemide), to manage pulmonary edema and lower preload
- Use catecholamines (dobutamine, adrenaline, NOT dopamine), preferably milrinone depending on blood pressure and myocardial function. (If there is a subvalvular stenosis, catecholamines should be administered with particular caution due to a possible increase in the obstruction.)
- Assess by ECHO and reduce afterload (e.g., sodium nitroprusside)
- Aim for moderate metabolic acidosis (target pH 7.35)
- Administer as little volume as possible; preferably only after cardiac function has recovered or is assessed to be stable in echocardiography

*1.6.1.1. Critical aortic stenosis (AoS)*

*1.6.1.1.1. Hemodynamic situation*

Due to the severe obstruction of the aortic valve, the left ventricular function usually is markedly impaired and cannot provide sufficient cardiac output for the systemic circulation which is supplied with blood from the pulmonary artery via the patent ductus arteriosus (i.e., prenatal situation). Left ventricular hypertrophy and possibly fibroelastosis have already developed in utero. This defect is often associated with other left heart obstructions (mitral stenosis, coarctation of the aorta, hypoplastic aortic arch—i.e., Shones complex). If a heart murmur is not already detected in the neonate, many patients with critical aortic stenosis present in severe cardiac shock. In addition, there is differential cyanosis.

In critical AoS with PDA-dependent systemic circulation (prostaglandin infusion), a left-to-right shunt at the atrial level is necessary to allow mixing of the oxygenated pulmonary venous blood via the right atrium, the right ventricle, the pulmonary artery and then the patent ductus arteriosus. A balloon atrial septostomy (Rashkind maneuver) may be necessary, if the shunt at the atrial level is not large enough,

*1.6.1.1.2. Further procedure*

If suspected ensure prompt transfer to a pediatric cardiac center for interventional catheterization and performance of balloon valvuloplasty (method of choice) or surgical commissurotomy (rarely required); possibly carry out an emergency Rashkind maneuver if there is a restrictive foramen ovale and inadequate left ventricular function after balloon valvuloplasty.

*1.6.1.2. Coarctation of the aorta (CoA)*

*1.6.1.2.1. Hemodynamic situation*

In patients with critical CoA, the lower half of the body is supplied with deoxygenated blood from the pulmonary artery via the PDA (differential cyanosis). When the ductus arteriosus closes, dramatic hypoperfusion of the body distal to the aortic isthmus occurs. The left ventricle has now suddenly contract against the pronounced obstruction and rapid decompensation occurs. Associations with other left heart obstructions (bicuspid aortic valve) or a VSD are common.

*1.6.1.2.2. Further procedure*

Again organize prompt transfer to a pediatric cardiac center for surgical correction, which is generally attempted after the hemodynamic situation has been stabilized. In some special conditions (e.g., for patients in poor general condition or with necrotizing enterocolitis—NEC), primary interventional catheterization with balloon dilatation or implantation of a coronary stent may be indicated for stabilization until surgery.

*1.6.1.3. Interrupted aortic arch*

*1.6.1.3.1. Hemodynamic situation*

There are different forms of an interrupted aortic arch and the perfusion of the lower half of the body depends however entirely on a patent ductus arteriosus (i.e., prostaglandin). A VSD is nearly always associated with this defect and other left heart obstructions occur occasionally. Again oxygen saturation should be measured preductally (right hand), as the preductal saturation reflects the situation of the perfusion and blood supply in the CNS and coronary arteries. The levels measured in the lower limbs correspond with pulmonary arterial saturation.

*1.6.1.3.2. Further procedure*

Prompt transfer to a pediatric cardiac center for surgical correction.

*1.6.1.4. Hypoplastic left heart syndrome*

*1.6.1.4.1. Hemodynamic situation*

In patients with a hypoplastic left heart syndrome (HLHS), both the pulmonary and systemic systems are supplied by the right ventricle. Although there are four forms of HLHS (i.e., MA

and AoA, MS and AoA, MS and AoS, MA and AoS with VSD), retrograde coronary perfusion takes place in all across the ductus arteriosus with mixed blood from the pulmonary artery and retrogradely across the coarctation and ascending aorta. Within the first few hours of life, the drop in pulmonary resistance together with the manifestation of a coarctation causes the blood from the pulmonary artery to flow primarily into the pulmonary circulation. Therefore, the systemic circulation—together with the coronary arteries—is increasingly less perfused. Severe heart failure develops, which if untreated usually results in shock. As a result, there is severe metabolic acidosis but due to pulmonary recirculation, the oxygen saturation is only moderately reduced. The higher the oxygen saturation is, the more the ratio of pulmonary to systemic perfusion changes are in favor of pulmonary over perfusion and systemic underperfusion. In addition, postnatal closure of the PDA enhances this fatal circulation with underperfusion of the systemic circulation and excessive pulmonary perfusion [9].

It is important to note that oxygenated pulmonary venous blood from the left atrium can only reach the systemic circulation across a sufficiently large shunt to the right atrium (see above).

*1.6.1.4.2. Initial treatment*

- Shock treatment, including intubation and ventilation (but avoid ventilation if possible, as long as the pH is balanced); hyperventilation must be avoided
- Prostaglandin E1: initial dosage 50–100 ng/kg/min
- Avoid administering oxygen (additionally reduces pulmonary resistance and thus systemic perfusion), target saturation 70–85%
- Administer some furosemide (1–2 mg/kg) to lower preload or for pulmonary edema
- Catecholamine treatment is often required, but should be administered with restraint (can increase the myocardial oxygen consumption), give milrinoneor dobutamine if needed, do not use dopamine
- Possibly reduce afterload (e.g., sodium nitroprusside)
- For severe acidosis, large amounts of NaBic are often required to manage acidosis (doses up to 10 ml/kgbw of NaBic are common!)
- Aim for moderate metabolic acidosis (target pH 7.35, avoid over-buffering)
- As little volume as possible should be administered; preferably only after cardiac function has recovered or is assessed to be stable in echocardiography

*1.6.1.4.3. Further procedure*

If the atrial defect is restrictive (leading symptom: severe cyanosis, oxygen saturation <65%, seriously ill child), pulmonary congestion has developed and an emergency balloon atrial septostomy (Rashkind maneuver) may be needed.

The patient should be transferred quickly to a pediatric cardiac center for surgery after stabilization (usually a Norwood procedure as the first step of three-stage Fontan palliation).

*1.6.2. Right ventricle outflow tract obstruction lesion (critical pulmonary stenosis and pulmonary atresia with intact ventricular septum or ventricular septal defect, tetralogy of fallot, tricuspid atresia with pulmonary stenosis and Ebstein anomaly)*

The initial treatment of this group of patients has some common features:

- Prostaglandin E1 infusion: initial dosage 50–100 ng/kg/min
- Initially more generous oxygen therapy (lowers pulmonary resistance)
- Initially more generous volume therapy (i.e., 20 ml/kg bw)
- Try to achieve mild metabolic alkalosis (use over-buffering) (pH 7.45–7.5)
- Possibly ventilation and mild hyperventilation ($pCO_2$ around 35 mmHg)
- Possibly use an increase in systemic resistance (noradrenaline)

*1.6.2.1. Critical pulmonary stenosis*

*1.6.2.1.1. Hemodynamic situation*

A critical pulmonary stenosis is a high-grade obstruction of the pulmonary valve with subsequent hypoxemia. As the right ventricle cannot drain into the pulmonary circulation, secondary hypoxia occurs due to a right-to-left shunt across the foramen ovale/ASD. The musculature of the RV is severely hypertrophic and the RV and tricuspid valve may sometimes be hypoplastic. To ensure oxygenation, the pulmonary circulation is supplied with blood from the aorta via the PDA.

*1.6.2.1.2. Further procedure*

After diagnosis and prostaglandin infusion organize transfer to a pediatric cardiac center for interventional catheter balloon valvuloplasty.

*1.6.2.2. Pulmonary atresia with intact ventricular septum*

*1.6.2.2.1. Hemodynamic situation*

In pulmonary atresia with intact ventricular septum, which can be judged as extreme form of pulmonary stenosis, the right ventricle cannot drain its blood normally to the PA. The majority of blood from the right ventricle either flows back into the right atrium due to tricuspid regurgitation. In addition in some cases, the right ventricle may be connected with the coronary arteries via myocardial sinusoids. In the latter case, the coronary arteries may have other problems and are stenotic so that coronary perfusion may depend on blood flow from the right ventricle (right ventricular dependent coronary circulation (RVDCC). Usually there is

suprasystemic pressure in the right ventricle and the right ventricle is hypoplastictic various degrees. Again, the pulmonary circulation is supplied with blood from the aorta via the PDA to ensure oxygenation.

*1.6.2.2.2. Further procedure*

After stabilization, organize transfer to a pediatric cardiac center. In most cases, cardiac catheterization is needed to rule out myocardial sinusoids and coronary anomalies. In some cases, it is possible to open the right ventricular outflow tract by interventional catheterization or implant a stent into the PDA. Otherwise interim palliative surgery (opening the right ventricular outflow tract or aortopulmonary shunt) must be performed accordingly.

*1.6.2.3. Tricuspid atresia*

*1.6.2.3.1. Hemodynamic situation*

In tricuspid atresia, that may give an example for all univentricular heart defects, there is no continuity between the right atrium and ventricle, so the right atrium can drain into the left atrium only across a right-to-left shunt at atrial level. There are various forms of tricuspid atresia depending on the variations of obstructions of the right ventricular outflow tract or pulmonic valve. The RV is usually perfused from the LV across a VSD that is almost always present. The RV is hypoplastic to various extents. Oxygen saturation is identical in the aorta and pulmonary artery because of the complete mixing of blood.

Frequently stenosis and even atresia of the pulmonary artery occur and may have a negative effect on the blood flow to the pulmonary circulation. If there is atresia or a high grade stenosis of the pulmonary artery, perfusion of the lungs depends on a patent ductus arteriosus (see above). In addition, the great vessels can be in normal position or in transposition.

If there is no pulmonary stenosis and blood flow to the pulmonary circulation is unobstructed, the main symptom that may develop later on is heart failure (tachypnea, hepatomegaly, pallor, possible pulmonary edema), but this constellation is much less common.

Initial treatment if there is no pulmonary stenosis (leading symptom: heart failure):

- Anticongestive treatment (diuretics, milrinone, rarely catecholamines)
- Restrictive volume therapy
- Restrictive oxygen therapy

*1.6.2.3.2. Further procedure*

Transfer to a pediatric cardiac center should be organized. If there is a restrictive atrial shunt (rare), an interventional catheter balloon atrial septostomy (Rashkind maneuver) may be needed. If there is inadequate pulmonary perfusion, a palliative aortopulmonary shunt is placed. The separation of circulations is performed later as multistep procedure (Fontan procedure).

#### *1.6.2.4. Tetralogy of Fallot*

##### *1.6.2.4.1. Hemodynamic situation*

In tetralogy of Fallot, there is a typical combination of a large VSD with overriding of the aorta, a subpulmonary and pulmonary stenosis, often combined with supravalvular stenosis of various degrees and a marked hypertrophy of the right ventricle. Cyanosis is determined by the extent of the obstruction of the right ventricular outflow tract and thereby pulmonary blood flow. In most cases, this obstruction is only mild at birth, but becomes more significant during the first few weeks of life due to the increase in the infundibular stenosis. If there is pronounced stenosis of the right ventricular outflow tract (functional pulmonary atresia), the children may already develop symptoms with cyanosis or hypoxic spells even in the neonatal period. Hypercyanotic spells occur if there is a sudden increase in subpulmonary muscular obstruction of the RVOT (i.e., agitation) or an acute drop in peripheral vascular resistance (i.e., after feeding).

Treatment of a hypoxic-hypercyanotic spell:

- Immediate sedation (e.g., ketamine IV 2–5 mg/kg, alternatively opiates, benzodiazepines)
- Oxygen therapy
- Increase in systemic resistance by
  - pressing the child's flexed knee against the chest ("jack-knife position")
  - if necessary infusion of vasoconstrictors (noradrenaline)
  - generous volume bolus (e.g., 20–50 ml/kg)
- Compensation of metabolic acidosis by buffering
- Possibly beta blockers (e.g., propranolol IV 0.01–0.1 mg/kg very slowly under monitor guidance)

##### *1.6.2.4.2. Further procedure*

Rapid transfer to a pediatric cardiac center is urgently needed. In most circumstances, interventional management can improve the hemodynamic situation (i.e., balloon dilatation of the pulmonary valve, stenting of the right ventricular outflow tract, stenting of a PDA). Only in few cases, early surgical correction is required today. In special situations (e.g., very small children), an aortopulmonary shunt is first placed as a palliative measure to ensure lung perfusion.

#### *1.6.2.5. Pulmonary atresia with ventricular septal defect*

##### *1.6.2.5.1. Hemodynamic situation*

From the hemodynamic aspect, this disease is an extreme form of Tetralogy of Fallot. Pulmonary perfusion is, however, dependent on a patent ductus arteriosus (prostaglandin infusion) or aortopulmonary collaterals.

*1.6.2.5.2. Further procedure*

After stabilization with prostaglandin infusion, a rapid transfer to a pediatric cardiac center should be organized. In cases with only membranous valvular atresia, an attempt can be made to open the valve by interventional catheterization (or with stent placement). In addition placing a stent in the PDA may help to achieve catch-up growth of the pulmonary vessels, which are almost always hypoplastic. If this intervention is not possible, an aortopulmonary shunt is placed as surgical palliation. After catch-up growth of the pulmonary vascular system, a continuity between the pulmonary vessels and the right ventricle is created surgically later (e.g., with a valved conduit).

*1.6.2.6. Ebstein's anomaly*

*1.6.2.6.1. Hemodynamic situation*

In Ebstein's anomaly, there is apical displacement of the tricuspid valve into the right ventricle combined with moderate to severe tricuspid regurgitation and reduced flow from the small right ventricle to the pulmonary artery. The right atrium is markedly too massively dilated. A pronounced Ebstein's anomaly can already be symptomatic in the neonatal period. Cyanosis occurs due to a right-to-left shunt at the atrial level caused by minimal antegrade flow to the pulmonary artery. In addition, the gross dilatation of the heart may compress the lungs and impair pulmonary function.

The disease may often complicated by accessory pathways (Wolf-Parkinson-White syndrome).

Besides initial treatment stated above, in Ebstein's anomaly:

- If there is heart failure, possibly catecholamines and diuretics
- If there is supraventricular tachycardia, it should be terminated with a vagal maneuver, adenosine, amiodarone, possibly cardioversion

*1.6.2.6.2. Further procedure*

The goal of surgical treatment is to reconstruct the tricuspid valve and close the ASD. If there is insufficient lung perfusion, it may be necessary to place an aortopulmonary shunt. If there is pronounced hypoplasia of the right ventricle, the univentricular Fontan pathway may be the only option. The overall prognosis is unfavorable for children who become symptomatic already in the neonatal period. These children require urgent transfer to a pediatric cardiac center.

*1.6.3. d-transposition of the great arteries (d-TGA)*

*1.6.3.1. Hemodynamic situation*

In d-TGA, there is parallel connection of the pulmonary and systemic circulations; the systemic venous blood is directed back into the aorta and the pulmonary venous blood is

pumped back to the pulmonary artery. Survival is only possible if shunts between the two circulatory systems do exist. The most important is a patent foramen ovale or ASD as shunt at atrial level, so oxygenated blood can reach the systemic circulation via the right ventricle across this left-to-right shunt. A PDA has a favorable effect on oxygenation because PDA blood flow increases lung perfusion and as a result the volume load and pressure in the left atrium is increased, so the left-to-right shunt at atrial level increases and more oxygenated blood can reach the systemic circulation [10].

Oxygen application may act indirectly by reducing pulmonary resistance and thus increasing pulmonary perfusion and thereby left atrial volume load and pressure (caution: uncontrolled administration of oxygen can also lead to closure of the ductus arteriosus when used without prostaglandin).

In addition to improve mixing between the two circulations, especially in "poor mixers," an attempt should be made to improve mixed venous saturation.

If there is an associated large VSD, cyanosis is usually less pronounced.

Initial treatment:

- Prostaglandin E1 infusion: initial dosage 50 ng/kg/min
- Generous volume therapy
- Aim for mild metabolic alkalosis (over-buffering) (pH 7.45–7.5, lowers pulmonary resistance)
- Oxygen therapy for severe cyanosis in neonates (caution: induces closure of the ductus arteriosus)
- If possible avoid intubation, ventilation and relaxation (lowers oxygen consumption and therefore increases mixed venous saturation. On the other hand, ventilation increases the intrathoracic pressure, which can impair mixing of the blood)
- Consider the improve cardiac output and thus mixed venous saturation by milrinone
- Generous treatment of anemia (improves oxygen supply)

*1.6.3.2. Further procedure*

If there is a restrictive shunt at the atrial level, a bedside interventional catheter balloon atrial septostomy (Rashkind maneuver) should be performed as soon as possible before transfer to a pediatric cardiac center. The surgical standard treatment is an arterial switch operation (Jatene procedure) within the first 2 weeks of life.

*1.6.4. Truncus arteriosus communis*

*1.6.4.1. Hemodynamic situation*

In a truncus arteriosus communis (TAC), only one vessel (common trunk) arises from the heart. The systemic and pulmonary circulation and the coronaries are here supplied from this

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trunk only. A VSD is almost always present. Since the blood follows the path of least resistance and tends to flow into the pulmonary circulation, there is usually excessive pulmonary blood flow after the drop in pulmonary resistance between the second and eighth weeks of life. In this situation, clinical signs of heart failure are already present in the first weeks of life. Due to the significant pulmonary recirculation, there is often only astonishingly mild cyanosis, although the trunk vessel contains only mixed blood.

Initial treatment:

- Oxygen lowers pulmonary resistance and thus increases blood flow to the pulmonary circulation, leading to excessive pulmonary blood flow and increasing heart failure—therefore, use oxygen restrictively
- Treatment of heart failure: diuretics, possibly catecholamines (dobutamine) or phosphodiesterase inhibitor (milrinone), afterload reducer (ACE inhibitor, possibly sodium nitroprusside)
- If associated with an interrupted aortic arch: prostaglandin E1 infusion: initial dosage 50–100 ng/kg/min

#### *1.6.4.2. Further procedure*

Organize transfer to a pediatric cardiac center. The definitive correction with a Rastelli procedure is generally performed within the first weeks of life depending on the clinical signs of heart failure.

### *1.6.5. Total anomalous pulmonary venous connection (TAPVC)*

#### *1.6.5.1. Hemodynamic situation*

Depending on the location of the anomalous connection of the pulmonary veins, we distinguish between supracardiac, cardiac, infracardiac and mixed forms. Please note that an infracardiac total anomalous pulmonary venous connection is regularly associated with an obstruction at the connection site.

In a total anomalous pulmonary venous connection, all pulmonary veins drain into the systemic venous system and oxygenated blood is guided from there into the right atrium. The perfusion of the systemic circulation thus depends on a right-to-left shunt at the atrial level that is necessary for survival.

In a TAPVC without obstruction of the pulmonary vein, the hemodynamic situation is similar to that of a large ASD (volume overload of the right atrium, ventricle and pulmonary circulation).

Initial treatment:

Total anomalous pulmonary venous connection with obstruction of the pulmonary vein:

- Oxygen therapy
- Intubation and ventilation with a high PEEP (>10 cm $H_2O$) for pulmonary edema

- Lower PVR: hyperventilation, generous buffering (target pH 7.45–7.5), increase oxygen supply, possibly inhaled NO or prostacyclin IV
- Diuretics, possibly catecholamines for low cardiac output (caution: catecholamines can exacerbate pulmonary edema)

*1.6.5.2. Further procedure*

A TAPVC with obstruction of the pulmonary vein is an absolute cardiac surgery emergency that must be surgically corrected immediately. Dilatation with or without stent implantation can treat the stenosis of the pulmonary vein in individual cases, so that the surgical repair can be performed after the child is stabilized.

If there is a restrictive atrial shunt or if surgical correction is not possible immediately, an interventional catheter balloon atrial septostomy (Rashkind maneuver) may be considered.

*1.6.6. Complete atrioventricular septal defect*

*1.6.6.1. Hemodynamic situation*

In a complete atrioventricular septal defect (AVSD, AV canal), both segments of the ventricular and atrial septum in the region of the AV valves are absent and the development of the AV valves is also impaired. This malformation results in a large left-to-right shunt that increases when the pulmonary resistance drops within the first weeks of life. Children with a complete AVSD usually develop heart failure when the pulmonary resistance drops after 2–8 weeks of life. The situation can be complicated by (mainly systemic) AV valve insufficiency.

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**Note**

trisomy 21 is frequently associated with AVSD. All neonates with trisomy 21 must therefore have an ECHO examination at an early age.

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Initial treatment:

- Avoid oxygen (excessive pulmonary blood flow is increased)
- Pharmacological treatment of heart failure: diuretics, ACE inhibitors, beta blockers, rarely catecholamines (possibly digoxin?)

*1.6.6.2. Further procedure*

The corrective surgery is generally performed at the age of 4–6 months and it may be necessary earlier if conservative heart failure treatment is unsuccessful.

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## 2. Postoperative cardiac intensive care therapy

The following remarks apply primarily to those patients that are transferred immediately after surgery from the surgical operating room and their early postoperative period in the pediatric cardiac intensive care unit (PCICU).

### 2.1. Basics

One of the main requirements for providing good quality of care in a PCICU is to understand that the job of the intensive care physician starts far before the operated child is admitted to the PCICU. Detailed information on the case of the child is required in advance before the day of admission. The PCICU intensivist must be aware of and understand the hemodynamics of the cardiac defect (i.e., check the ECHO, cath data, etc.), possible complicating or concomitant diseases, the actual planned surgical repair or procedure and the usually possible perioperative and postoperative complications.

Every intensive care physician should be familiar with the major steps of cardiac surgery in general and the specific steps and details of the case to be admitted. These steps include the whole process of surgery from opening the thorax and the mediastinum, thereafter arterial and venous cannulation for the cardiopulmonary bypass, initiation of cardiopulmonary bypass, cardioplegic arrest and finally, the restoration of cardiac function at the end of the operation including decannulation and closure of the thorax.

To obtain this crucial understanding, every intensivist should visit the cardiac theatre on a regular basis and observe about 50 surgeries per year. In complex cases, the intensivist should work together in the time after decannulation or weaning of the bypass with the anesthetist before admission of the patient to the PCICU.

### 2.2. Preparation of the bed place

The majority of patients will be admitted on a planned and forseable basis. Before the patient is transferred to the PCICU, a careful and standardized preparation of the bed/place must be completed to avoid unnecessary misunderstanding and streamline the admission process. This preparation may vary from unit to unit but generally includes the following:

- Preparation of the ventilation equipment, adjustment of the settings to the estimated patient-specific parameters, checking of the ventilation bag and mask, oxygen delivery, suction equipment and catheters. In addition, it must be determined by contacting the operating theatre or by assessment of the preoperative information whether iNO (nitric oxide) ventilation is required and possible with the ventilation equipment
- Medication plan: The prepared medication plan should be tailored to the patients' age and weight and include the medication and calculated dosages for the presumed standardized postoperative treatment (i.e., sedation, pain therapy, antibiotics, catecholamines, vasodilators, diuretics, fluid therapy and infusions). The perfusors and flushing solutions should be purged and primed before the patient is transferred to the PCICU

- Monitoring: The bedside monitor is checked beforehand and the patient-specific alarms according to reference values for age and disease are set
- Administrative tasks: Unit specific paperwork is prepared and an X-ray order for the first postoperative X-ray and laboratory order for the first laboratory tests should be prepared

### 2.3. Postoperative transfer of the patient to the ICU

In order to understand the physiology of the patient, a detailed handover from the different disciplines treating the patient during surgery is necessary. The checklist in **Table 2** contains some common and useful questions that the intensive care physician should discuss with the surgeons, anesthesiologists and cardiac technicians/perfusionists at transfer. The information is crucial and may have direct impact on the subsequent PCICU management.

| **Surgical aspects** |
|---|
| - What were the intraoperative findings? Compared to before? |
| - What surgical technique was performed? Any problems? |
| - What drainages were placed intraoperatively (e.g., pleural drainage, mediastinal drainage)? |
| - What is the assessment of postoperative result? |
| - Was an intraoperative transesophageal echocardiography performed and what were the findings (residual gradient, valve insufficiency, residual shunt, myocardial function)? |
| - Did any intraoperative arrhythmias occur? How were they treated? |
| - Did any intraoperative bleeding problems occur? How were they treated? |
| **Anesthesiology aspects** |
| - Tube size and brand, cuffed and uncuffed? |
| - Ventilation settings ($FiO_2$, tidal volume, rate, peak pressure and PEEP) |
| - Central venous catheter: location of insertion, size |
| - Arterial access: location, size |
| - Was an LA or pulmonary artery catheter placed? |
| - Anesthetics and cardiac medication used (cathecolamines, vasodilators, anti arrhythmics) and their dosages |
| - Heparinization and current coagulation levels |
| - Use of blood products (pack red cells, platelet concentrates, fresh frozen plasma) |
| **Cardiopulmonary bypass** |
| - Bypass time |
| - Aortic cross clamp time |
| - Cardiac arrest time |
| - Minimum temperature during bypass |
| - Was hemofiltration performed at the end of the bypass? |

**Table 2.** Checklist for postoperative transfer.

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### 2.4. Initial examination after transfer to the ICU

Immediately after the patient is admitted to the ICU, the most important clinical parameters must be assessed by the admitting physician and the nurses involved to confirm that the patient is in a stable and safe situation:

- Respiration: Is the chest moving? How are thorax/chest movements? Symmetrical lung expansion? Adequate expansion? How is the Oxygen saturation? FiO2?
- Circulation: Palpation of peripheral pulses (femoral, brachial), visual check of pressure parameters on the monitor (arterial blood pressure, CVP, if inserted pulmonary artery pressure or LA pressure) and check of capillary filling time (normal<2–3 s)
- Diuresis: Is the urine bag filled? How much? Is the bladder filled despite urine catheter? Does the urine look clear or hemolytic? Does the patient appear edematous (puffy) or rather dehydrated?
- Bleeding: Are the drains connected to suction? How much is in the bag and how quick is it filling? Are the secretions dark red (venous blood), bright red (arterial blood), yellowish or clear (serous), warm (fresh) or cold (older)?
- Neurology: is the patient sedated/anesthetized, what is the status of the pupils, are there spontaneous movements and is the patient breathing against the ventilator?
- Body temperature
- Assessment of catheters, lines and cables inserted

During this gross examination to assess the patient's stability, the patient is connected to the bedside monitor system and other equipment. The arterial and central venous accesses are connected with the pressure transducers, set to zero and activated again. The patient is also connected to the ventilator and the drainages set to suction (in general, the suction pressure is set at around 15 cm $H_2O$). In addition, the external pacemaker is checked for proper function.

In parallel, the first blood samples are taken and include an arterial and central venous blood gas analysis, an ACT and a survey of different pathology markers to asses organ function. The first blood gas analysis including electrolyte status provides information on the ventilation situation, the hemodynamic status (lactate, central venous saturation) and electrolyte metabolism (especially potassium). A standard limb ECG is then recorded, a chest X-ray arranged and finally, an initial exploratory echocardiography (ECHO) performed as a standard in every patient.

### 2.5. Further postoperative management

#### *2.5.1. Fluid intake*

As capillary leak is a normal reaction to bypass and the subsequent inflammatory process induces edema of all organs, the amount of free water intake should be limited postoperatively. Therefore, fluid intake is ideally reduced to 30–50% of the normal maintenance on the first postoperative day and then increased day by day to 75 and finally 100% on the following days.

Patients, however, who underwent cardiac surgery without cardiopulmonary bypass (e.g., resection of an aortic coarctation, PDA ligature) do not require any restriction of postoperative fluid intake.

As soon as the children are able to drink, oral fluid intake should not be restricted any more.

*2.5.2. Ventilation*

The majority of patients are ventilated after cardiac surgery when they arrive in the PCICU. Pulmonary function is affected by a large variety of preoperative, intraoperative and postoperative factors. Important examples to be considered are listed below:

- Preoperative factors:
  - Preexisting excessive pulmonary blood flow (wet lung, congestion)
  - Preexisting pulmonary hypertension (left-to-right shunt)
  - External compression of the airways, for example, due to vascular rings or prominent pulmonary arteries
- Intraoperative factors:
  - Atelectasis due to surgical manipulation in the thorax or lack of ventilation during the bypass
  - Edema development due to the effect of cardiopulmonary bypass and SIRS
    - Trauma of the lung
- Postoperative factors:
  - Swollen mucosa (SIRS)
  - Atelectasis
  - Lung edema as a result of a postoperative increased lung perfusion. Typical examples are after a shunt procedure or operative opening of circulation in the lungs in pulmonary atresia or severe stenosis ("reperfusion edema")
  - Impaired respiratory mechanics due to postoperative diaphragmatic paresis
  - Pain-related reduction in respiratory capacity (surgery drains)
  - Respiratory depression caused by drugs
  - Pneumothorax, pleural effusions
    - Bleeding through MAPCAS

*2.5.3. Settings of the ventilator*

Volume-controlled ventilation is the ventilation of choice in every cardiac patient after cardiac surgery. In many ventilators, a setting with a pressure control combination (i.e., PRVC-mode)

can be chosen. Hereby markedly, lower rates are usually used (e.g., 20/min for neonates) in the sedated patient. The tidal volume is usually set to 10–15 ml/kg. A relatively long inspiration time is usually selected initially. The PEEP is around 5–10 mmHg and an $FiO_2$ value is selected at which oxygen saturation of over 95% can be achieved. When the patients waking up, the volume controlled mode is combine with a SIMV mode.

In spontaneous breathing patients, the following values can be used as references for the age-specific respiratory rate:

- Neonates: 40/min
- Infants: 25–30/min
- Toddlers: 25/min
- School-age children: 20/min
- Adolescents: 15/min

Again, in the controlled situation, the rate settings used are much slower.

The following situations are important variants from these initial settings of the ventilator:

- *Univentricular heart*: In univentricular hearts, the balance between systemic and pulmonary circulation is very important to avoid pulmonary overcirculation at the cost of inadequate systemic perfusion. At an oxygen saturation of around 75–85%, a nice balance between systemic and pulmonary perfusion is achieved (Qp/Qs = 1:1). Therefore, the target oxygen saturation in these patients is approximately 80%. Uncritical oxygen administration must be avoided.
- *Fontan circulation*: In these patients, the transpulmonary blood flow is depending completely on the intrathoracic pressure and pulmonary vascular resistance. Theoretically, a high PEEP increases the intrathoracic pressure and thus reduces passive blood across the pulmonary vascular system. A lower PEEP level (<5 mmHg) is therefore often recommended for Fontan patients. On the other hand, a higher PEEP also increases the residual functional capacity and reduces atelectases, so better lung perfusion can be achieved via the Euler–Liljestrand reflex. It has been advocated earlier to ventilate Fontan patients without PEEP—this management strategy is inappropriate and no longer recommended.
- *Pulmonary hypertension*: Pulmonary vascular resistance is reduced by a low $pCO_2$, a high pH and a high $FiO_2$. To use this principle, patients with known or presumed pulmonary hypertension (i.e., large VSD, older age, truncus, etc.) should be ventilated using mild hyperventilation (Target $pCO_2$: 30–35 mmHg) and with higher than normal $FiO_2$ to achieve an oxygen saturation of around 100% and a $paO_2$ of more than 150 mmHg

*2.5.4. Infection prophylaxis*

Cardiopulmonary bypass reduces the capacity of the immune system to act against bacterial infections and so postoperative antibiotic prophylaxis is always given for children after cardiac surgery. The antibiotics used and the duration of antibiotic treatment are, however, subject of some controversy. First or second generation cephalosporins are frequently used,

for example, cefazolin 30 mg/kg/dose every 8 hourly. For an uncomplicated postoperative course, many centers carry out antibiotic prophylaxis for 2–3 days (6–9 doses, until the drains are out) although hardly any evidence-based data for this procedure are available. For patients with an open sternum or peritoneal dialysis or ECMO, no additional management is required but prolonged treatment is often used in many centers. Antibiotic prophylaxis is then continued for 24 h post-chest closure or ECMO/PD removal.

#### *2.5.5. Sedation*

In most centers, a combination of an opiate (morphine, fentanyl) and a benzodiazepine (midazolam) is administered intravenously continuously in the initial postoperative period. Propofol is often used in older children who are sedated for only a short period. No time (2–3 days) or dose (3–8 mg/kg/h) limitation applies in these cases. Lactate levels should be checked on a routine basis.

#### *2.5.6. Analgesia*

Surgery itself and the drains in the body after surgery is painful procedure. To reduce postoperative pain, a fixed and standardized combination of non-steroidal analgesics (i.e., paracetamol, ibuprofen) and opiates (morphine, fentanyl) is usually used for analgesia for the first 2–3 days. Patient-controlled analgesia with a PCA pump is also applicable for older children (who can already play a video game).

#### *2.5.7. Kidney function*

Kidney function nearly always deteriorates in patients who have undergone cardiac surgery. The use of cardiopulmonary bypass leads to renal impairment and intraoperative fluid loading and inflammatory reactions that result in fluid retention. A negative fluid balance is therefore targeted postoperatively.

Urine excretion reflects a combination of cardiac function and cardiac output in combination with kidney function. In general, a urine excretion of at least 2 ml/kg/h after cardiac surgery is a minimum to ensure adequate fluid balance. Patients most often have a combination of pre-renal kidney failure (low cardiac output, capillary leak, volume deficit) in combination with the direct effects of SIRS on the kidney itself. In addition, a low systemic blood pressure and, in case of ascites, a high intra-abdominal pressure leads to a low perfusion pressure in the kidneys (aggressive drainage of ascites). Other less common examples of renal kidney failure are renal vein thrombosis or iatrogenic kidney damage (aminoglycosides, cyclosporin A). Post-renal kidney failure can be caused by obstructions in the region of the efferent urinary tract (e.g., obstruction of the urinary catheter or the urethra) or simply by increased intra abdominal pressure caused by capillary leak and ascites.

Suitable diuretics in the postoperative phase are primarily loop diuretics (Furosemide, ethacrynic acid). They are administered as a bolus or as a continuous infusion if the response is inadequate. The combination with theophylline sometimes enhances the effect. If excretion is insufficient or the fluid balance is clearly positive, peritoneal fluid drainage, or peritoneal dialysis should be initiated early.

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### 2.6. Common postoperative problems and complications

Typical postoperative problems and their most common causes are summarized in **Table 3**.

| Problem | Common causes |
|---|---|
| Blood pressure too high | Pain, fear, catecholamines, excessive fluid volume, result of the abrupt drug discontinuation (beta blockers, ACE inhibitors);<br>typical postoperative problem after correction of an aortic coarctation.<br>Rare causes: cerebral seizures, hypoglycemia (counter regulation) |
| Blood pressure too low | Hypovolemia, low cardiac output: limited myocardial function, pericardial effusion, arrhythmia (junctional ectopic tachycardia, AV block), excessive drainage loss, hemorrhage, excessive diuresis; vasodilators, anaphylaxis, sepsis, shock, pneumothorax |
| Central venous pressure (CVP) too low | Fluid deficit (excessive drainage losses, hemorrhage, excessive diuresis, volume intake too low) |
| Central venous pressure (CVP) too high | Tense/stiff patient (reaction to wake up, insufficient sedation in ventilation patients); impaired right ventricle function. In patients with univentricular heart, a high CVP suggests poor systemic ventricular or a relevant AV valve regurgitation. Other causes are a pericardial tamponade or pneumothorax |
| Arterial saturation too low | Atelectasis, hypoventilation, technical problems with the ventilation device, disconnected/obstructed tube, pneumothorax, pleural effusion, pneumonia, pulmonary edema, pulmonary hemorrhage, secretion, right to left shunt |
| Arterial saturation too high | In patients with univentricular hearts, saturation over 85% suggest an imbalance between pulmonary and systemic perfusion: excessive blood flow to the lungs and diminished supply to the systemic circulation |
| Bradycardia | Sinus bradycardia, AV block |
| Tachycardia | Narrow QRS complexes: sinus tachycardia, supraventricular tachycardia, JET<br>Wide QRS complexes: ventricular tachycardia |
| Increase in lactate | Poor systemic perfusion, seizures, gut ischemia |

**Table 3.** Typical postoperative problems and the most common causes.

#### *2.6.1. Low cardiac output syndrome (LCOS)*

There is a variety of factors that can result in myocardial dysfunction and subsequent low cardiac output postoperatively. Typically, they include an inflammatory reaction to cardiopulmonary bypass, myocardial ischemia and inadequate myocardial function as a result of the intraoperative clamping of the aorta, intraoperative hypothermia, a reperfusion edema, or if a surgical procedure was performed using a ventriculotomy—direct myocardial damage, coronary ischemia, inadequate cardioplegia, mechanical alteration, or an infection.

##### *2.6.1.1. Symptoms*

Typical clinical signs of low cardiac output are as follows:

- Tachycardia
- Oliguria

- Delayed capillary filling time (>3–4 s)
- Hypotension
- Decreased pulse pressure
- Reduced mixed venous saturation (Note: a difference between arterial and mixed venous saturation not <20–25% suggests sufficient cardiac output and adequate oxygen supply)
- Metabolic acidosis (BE less than −5)
- High lactate level (>3 mmol/l)

*2.6.1.2. Treatment*

The treatment of low cardiac output has to focus on the underlying causes which should be eliminated if possible. The following measures depending on the hemodynamic situation are commonly used as follows:

- Volume substitution (in case of hypovolemia)
- Inotropic support (catecholamines (i.e., adrenaline, dobutamine), phosphodiesterase inhibitors, intravenous calcium)
- Afterload reduction (sodium nitroprusside, phosphodiesterase inhibitors)
- Consider hormonal therapy for hypotensive resistance to epinephrine and vasopressors: cortisol for suspected adrenal insufficiency, IV or oral triidothyronine (T3) for euthyroid sick syndrome (low T3 levels with LCOS symptoms)
- Chronotropic support (pacemaker therapy, positive chronotropic drugs)
- Treatment of arrhythmias (amiodarone)
- Ventilation strategy
- Reduction in oxygen consumption (sedation, cooling)
- Mechanical circulatory support (ECMO, LVAD)

*2.6.2. Pulmonary hypertensive crisis*

Based on the underlying anatomy and pathophysiology, many patients may have a significant elevation of pulmonary pressures or pulmonary vascular resistance before surgery. Typical heart defects for this are those with high flow and pressure driven left-to-right shunt defects (large VSD, AVSD, Truncus, etc.) and those with unrestricted flow over a long period (i.e., >6 or 12 months of age). Postoperatively, there may be a rapid and critical increase in pulmonary arterial pressure or pulmonary vascular resistance in certain situations (i.e., agitation, external stimulation such as suctioning, pain, etc.). The result of a rapid increase in PVR is a standstill of the trans-pulmonary blood flow with secondary congestion in the right atrium (high CVP) and ventricle and drop in pressure in the left atrium and ventricle (low CO and BP). Ultimately, cardiac output collapses and the coronaries are not perfused. The following patients are at a particularly high risk for this type of crisis:

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- Patients with already increased pulmonary vascular resistance preoperatively (primary pulmonary hypertension)
- Neonates within the first week of life
- Patients with pulmonary venous hypertension (e.g., within the context of a total anomalous pulmonary venous connection or mitral stenosis)
- Older children with a still uncorrected high flow and pressure shunt defects that led to an increase in pulmonary vascular resistance (e.g., complete AV canal, large VSD, truncus)

The following factors may increase pulmonary vascular resistance:

- Hypoxia
- Acidosis (pH <7.3, BE less than −5)
- High partial pressure of carbon dioxide ($paCO_2$ >50)
- Polycythemia
- Atelectasis
- Agitation

The following factors may reduce pulmonary vascular resistance:

- Oxygen administration
- Alkalosis
- Hyperventilation
- NO inhalation (nitric oxide)
- Recruitment of atelectatic lung segments

Prophylaxis of a pulmonary hypertensive crisis is crucial, the acute management as well as the prophylaxis of a pulmonary hypertensive crisis includes the following measures:

- Sufficient analgesia and sedation or relaxation for 1–2 days
- Oxygenation ($paO_2$ >150)
- Optimizing the ventilation (adequate PEEP)
- Slightly alkalotic pH level (target pH 7.4–7.5) and mild hyperventilation ($paCo_2$ 30–35)
- Pharmacological vasodilators (NO, iloprost, prostacyclin)
- Avoidance of unnecessary manipulations such as too frequent suctioning

### 2.7. Specific early postoperative problems

Cardiac defects or surgery-specific postoperative problems and complications are summarized in **Table 4**.

| Cardiac defect/operation | Specific early postoperative problems and complications |
|---|---|
| ASD closure | Sinus node dysfunction, left heart failure/pulmonary edema in older children and adults |
| VSD closure | Pulmonary hypertensive crisis, complete AV block, JET, residual shunt |
| AV canal correction | Pulmonary hypertensive crisis, complete AV block, JET, AV valve stenosis or incompetence |
| PDA ligation | Injury to the recurrent laryngeal nerve (vocal cord paralysis) or the thoracic duct (chylothorax), accidental ligation or injury to surrounding vessels (especially left pulmonary artery, aorta |
| Truncus arterious correction | Pulmonary hypertensive crisis, truncus valve stenosis or incompetence, right ventricular dysfunction |
| Aortopulmonary window (correction) | Pulmonary hypertensive crisis, coronary ischemia |
| Anomalous pulmonary venous connection (correction) | Pulmonary hypertensive crisis, atrial arrhythmia, residual stenosis of the pulmonary veins or pulmonary vein anastomosis with the left ventricle, high left ventricular filling pressure due to the relatively small left atrium and ventricle |
| Fallot correction | Right ventricular diastolic dysfunction (poor compliance of the hypertrophic ventricle), JET, complete AV block, residual pulmonary stenosis, residual VSD, pulmonary insufficiency after a transannular patch |
| Pulmonary atresia with intact ventricular septum | Right ventricular dysfunction, myocardial ischemia due to right ventricle dependent coronary circulation, circular shunting due to creation of an aortopulmonary shunt and opening of right ventricular outflow tract |
| Aortic stenosis correction | Residual stenosis, disruption of left ventricular diastolic function, aortic insufficiency, AV block |
| Ross procedure | Coronary ischemia |
| Konno procedure | Coronary ischemia, obstruction of the right ventricular outflow tract, arrhythmias (e.g., AV block), mitral regurgitation |
| Subaortic stenosis (resection) | Residual stenosis, mitral valve injury, (ventricular) arrhythmia, AV block |
| Coarctation of the aorta (resection) | Residual obstruction, paraplegia, post-coarctectomy syndrome, unmasking an aortic valve stenosis, injury to the recurrent laryngeal nerve, chylothorax |
| Interrupted aortic arch (correction) | Residual obstruction, compression of the left main bronchus by the aorta, injury to the recurrent laryngeal nerve, chylothorax |
| Mitral stenosis correction | Pulmonary hypertensive crisis, residual stenosis, mitral regurgitation, left ventricular dysfunction |
| Creation of an aortopulmonary shunt | Imbalance between systemic and pulmonary perfusion, shunt leak, shunt thrombosis, pulmonary edema, coronary ischemia |
| Pulmonary artery banding | Cyanosis, insufficient banding (excessive pulmonary blood flow), Qp-Qs mismatch |
| Norwood procedure | Low cardiac output, imbalance between systemic and pulmonary perfusion, residual obstruction of the aortic arch, AV valve insufficiency, SIRS |
| Superior cavopulmonary anastomosis (Glenn, Hemifontan) | Cyanosis, hypertension, edema/congestion of the upper half of the body, Chylothorax |
| Fontan completion | Ascites, pleural effusions, edema, cyanosis, low cardiac output, arrhythmias |

| Cardiac defect/operation | Specific early postoperative problems and complications |
|---|---|
| TGA (switch operation) | Coronary ischemia, left ventricular dysfunction, neo aortic insufficiency, peripheral pulmonary stenosis |
| TGA (Mustard/Senning atrial baffle procedure) | Pulmonary or systemic venous obstruction, atrial arrhythmias |
| Bland-White-Garland syndrome (correction) | Myocardial dysfunction, mitral regurgitation |

**Table 4.** Specific early postoperative problem and complications (modified from Schwartz and Millar 2009 in Roger's Handbook of Pediatric Intensive care).

## 2.8. Postoperative features of heart defects with a univentricular heart

The large group of heart defects with a univentricular physiology may pose great challenges to postoperative intensive care in the PCICU. These patients include those with a hypoplastic left heart syndrome, tricuspid atresia, or a double inlet left ventricle. The most important principles in the postoperative treatment of these patients are presented below. As an example, the three-stage surgical procedure for patient with a hypoplastic left heart syndrome is explained. In general, palliation in a Fontan procedure is made with the goal of achieving complete separation of the pulmonary and systemic circulation to eliminate cyanosis. The lungs are perfused passively from the vena cavae without any pumping chamber in between. The single ventricle supplies only the systemic circulation and thereby has a reduced volume load.

The three stages are as follows:

- Norwood procedure or Damus-Kaye-Stansel procedure with a shunt
- Upper cavopulmonary anastomosis (Glenn procedure, "hemi Fontan")
- Total cavopulmonary anastomosis (Fontan procedure)

### *2.8.1. Norwood procedure*

The Norwood procedure is the first step for patients with a hypoplastic left heart syndrome toward separation of the circulatory systems by a Fontan procedure. There are several modifications of this surgical procedure (classical Norwood, Norwood-Sano-procedure). The principal goal is to form a neo-aorta from the pulmonary artery and the hypoplastic aorta that can supply the systemic circulation with blood without a pressure gradient (i.e., unobstructed systemic blood flow). To achieve this, the pulmonary artery and the hypoplastic aorta are anastomosed distal to the valves. Additional patch material is usually required for the reconstruction of the aortic arch and excision of the coarctation. In this manner, a strong vessel for systemic perfusion is created. The pulmonary artery is transected shortly before the pulmonary artery bifurcation and in most cases, pulmonary perfusion is ensured via an aortopulmonary shunt or Sano shunt (placement of a conduit through right ventricular to pulmonary artery).

To achieve the unobstructed inflow to the heart and outflow from the pulmonary veins, an atrial septectomy is also performed. There is a balance between the pulmonary and systemic circulatory systems when arterial saturation is between 75 and 85% (Qp/Qs = 1).

Typical problems after a Norwood procedure are low cardiac output, capillary leak caused by SIRS and hypoxemia.

*2.8.1.1. Low cardiac output*

As a typical result of the long bypass time required or even prolonged circulatory arrest, a systemic inflammatory response syndrome (SIRS) of various degrees occurs in these neonates. As a consequence, myocardial function is usually markedly impaired so that a certain and often higher amount of catecholamine support is always needed postoperatively. Other possible causes of inadequate systemic perfusion are increased pulmonary perfusion at the expense of systemic perfusion (Qp/Qs >1; leading symptoms: arterial saturation>85%, tachycardia, hypotension, oliguria, metabolic acidosis). To manage this situation, an attempt is made by reducing the afterload of the systemic circulation and increasing pulmonary resistance. AV valve insufficiency or arrhythmias can also cause or deteriorate low cardiac output.

*2.8.1.2. Hypoxemia*

Hypoxemia (i.e., saturation <70%) can be the result of an imbalance between pulmonary and systemic circulations to the detriment of the pulmonary circulation—for example, in an obstruction of the aortopulmonary shunt or increased pulmonary resistance.

Other causes are typical pulmonary problems such as atelectasis, a pleural effusion, edema, or pneumonia. Peripheral cyanosis occurs with low cardiac output or increased oxygen consumption (leading symptom: reduced systemic venous saturation).

*2.8.2. Superior cavopulmonary anastomosis*

The aim of a superior cavopulmonary anastomosis is to allow passive blood flow from the upper half of the body into the pulmonary circulation. The superior vena cava is anastomosed in an end-to side manor with the pulmonary artery and thereby the systemic venous blood from the upper half of the body then flows passively to the lungs without an intermediate pumping chamber and is oxygenated there. The systemic venous blood from the lower half of the body does not reach the lungs, but is mixed with the pulmonary venous blood in the heart and pumped into the systemic circulation. The systemic circulation thus contains mixed blood. The saturation of these children is usually about 80–85%.

This step leads to a complete hemodynamic unloading of the univentricular heart as the pulmonary circulation and systemic circulation are now connected in series. The Qp/Qs ratio is then 0.6–0.7; oxygen saturation is 80–85%. In comparison with older children, the head and upper limbs in young children are relatively large and therefore, the overall ratio of pulmonary perfusion is still higher in younger children. The percentage of systemic venous blood

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from the upper half of the body, that reaches the lungs via the cavopulmonary anastomosis and is oxygenated there, is correspondingly higher.

Typical postoperative problems are increased pressure in the superior vena cava, hypertension and hypoxemia.

*2.8.2.1. Elevated pressure in the superior vena cava*

In a superior cavopulmonary anastomosis, elevated pressure in the superior vena cava suggests an obstruction in the area of the cavopulmonary anastomosis or pulmonary circulation or elevated pulmonary vascular resistance.

The difference in pressure between the superior vena cava and atrium (transpulmonary gradient) should be <10 mmHg. A high pressure in the superior vena cava can restrict cerebral outflow and lead to marked edema in the upper half of the body (superior vena cava syndrome).

After this operation, patients should be positioned with the upper body elevated (about 45°, half sitting position). Elevated intrathoracic pressure from mechanical ventilation additionally hinders passive blood flow from the superior vena cava into the pulmonary circulation and patients should therefore be quickly extubated.

*2.8.2.2. Hypertension*

Temporary hypertension during the first few postoperative days is not unusual in these patients. The elevation of intracranial pressure that is necessary to maintain adequate cerebral perfusion pressure may be one reason. Aggressive lowering of the blood pressure should therefore be avoided but normal arterial pressures should be obtained.

*2.8.2.3. Hypoxemia*

Saturation levels below 75% following a superior cavopulmonary anastomosis can be caused by many reasons all resulting in a reduction in pulmonary perfusion. This may be due to an obstruction in the area of the anastomosis or pulmonary vessels. Another possible explanation is that blood from the upper half of the body is conducted past the alveoli, for example, if there are venovenous collaterals (connections between the systemic and pulmonary veins), the azygos vein was not ligated by the surgeon, a left sided superior vena cava has reopened or arteriovenous collaterals (connections between the pulmonary arteries and veins) have gained importance.

In the early postoperative phase, arterial saturation can often be improved by attempting mild hypoventilation. Slightly elevated $pCO_2$ (>50 mmHg) causes vasodilatation of the cerebral vessels, so more blood flows to the brain. Since this allows relatively more blood to reach the upper half of the body, more blood is conducted through the SVC and into the lungs and becomes oxygenated. In addition, the blood can also be slightly alkalized by administering sodium bicarbonate (target pH >7.4).

*2.8.3. Total cavopulmonary anastomosis (Fontan procedure)*

In a Fontan procedure (total cavopulmonary anastomosis), the pulmonary and systemic circulation are finally completely separated by anastomosing the inferior vena cava also with the pulmonary circulation. A tunnel is created that connects the inferior vena cava with the pulmonary artery. This tunnel may pass either through the atrium (intracardiac Fontan) or alternatively outside the heart (extracardiac tunnel). It should be standard to leave a small shunt between the Fontan tunnel and the atrium (i.e., Fontan fenestration) that functions as an overflow (right-to-left shunt) if the resistance in the pulmonary circulation is too high and not all of the systemic venous blood can enter the pulmonary circulation. This may have the disadvantage of mild hypoxemia but the positive effects (improved systemic circulation, decompression of the venous side, less ascites, edema, etc.) clearly overweigh the disadvantage of lower saturations.

When the Fontan circulation is completed, all of the systemic venous blood (exception: coronary sinus) flows passively into the pulmonary circulation without passing through a pumping ventricle. The oxygenated blood is then pumped into the systemic circulation by the univentricular heart.

Typical postoperative problems are low cardiac output, hypoxemia, effusions, arrhythmias and thrombosis.

*2.8.3.1. Low cardiac output*

In Fontan patients, low cardiac output can often occur due to low preload (hypovolemia), increased pulmonary resistance, or an obstruction in the area of the systemic venous outflow (tunnel stenosis, anastomosis stenosis). Therefore, higher volume requirements are often necessary in these patients. In addition, poor ventricular function or AV valve insufficiency and arrhythmias can also cause low cardiac output.

*2.8.3.2. Arrhythmias*

As the completion of the Fontan procedure require surgical manipulation at the atrium and the area of the sinus node, atrial arrhythmias may occur and sinus node dysfunction is typical. A pacemaker (atrial stimulation) sometimes becomes necessary.

*2.8.3.3. Hypoxemia*

Cyanosis can be the result of a relevant right-to-left shunt across a tunnel fenestration. In addition, the usual pulmonary problems (i.e., pleural effusion, atelectasis, pneumonia) can also lead to hypoxemia. Reduced pulmonary perfusion, caused by arteriovenous or venovenous collaterals can also cause hypoxemia in Fontan patients.

*2.8.3.4. Effusions*

Pleural effusions and ascites are common immediately after surgery. They can be the result of elevated venous pressure and volume load required and may lead to considerable postoperative complications.

*2.8.3.5. Thrombosis*

Based on the low flow in the Fontan circuit, these patients are at an increased risk of developing venous thrombosis. This risk is increased especially if there is low cardiac output. Most centers therefore recommend lifelong anticoagulation for Fontan patients. There is, however, no uniform opinion with respect to the duration or the form of anticoagulation (vitamin K antagonists or platelet aggregation inhibitors). While many different anticoagulation regimens are used in small children (no anticoagulation, aspirin, warfarin), there is a general consensus that anti-coagulation is obligatory in patients after puberty.

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Parts of this book chapter are similar to a recent publication of the third author [11]. Based on the content of the scientific information, this is, however, current state-of-the-art knowledge and medical management; therefore, similarities in the text are logical necessity.

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